

**Fig. 1a Non-Transduced hMSCs in Immuno-competent
mouse muscle
Day 3**

Dil + hMSCs

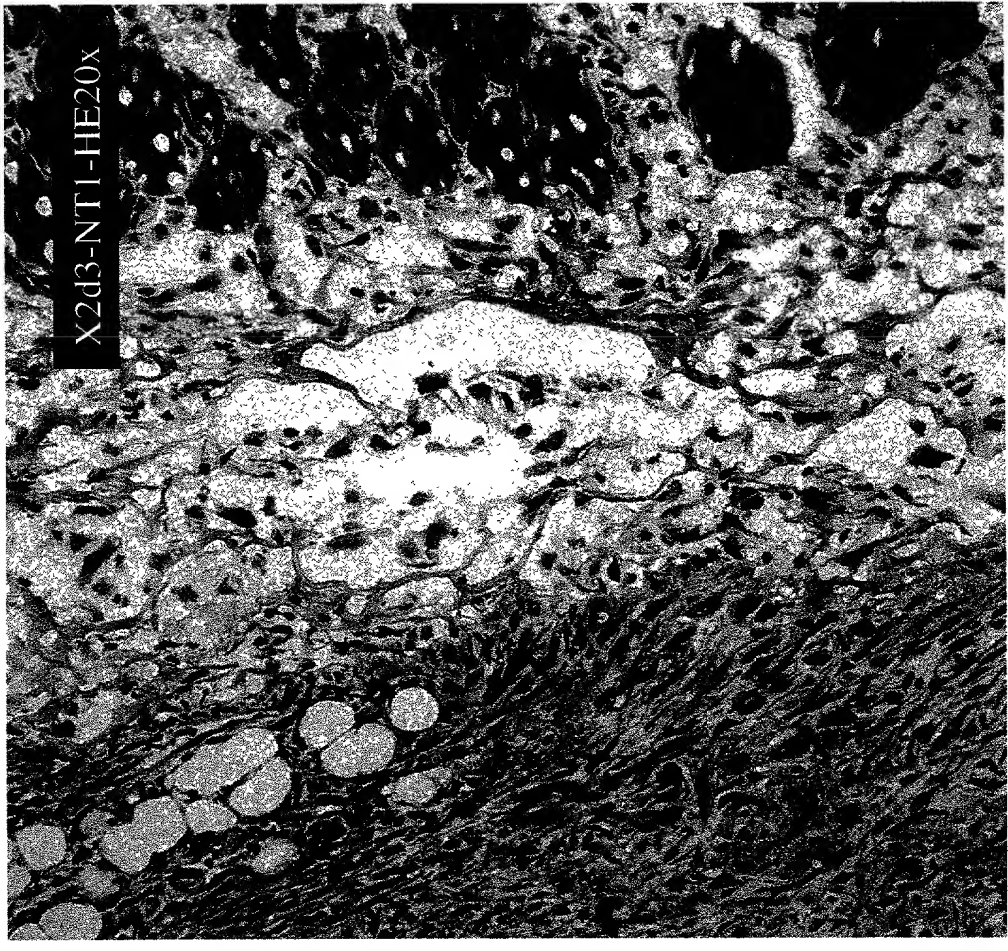
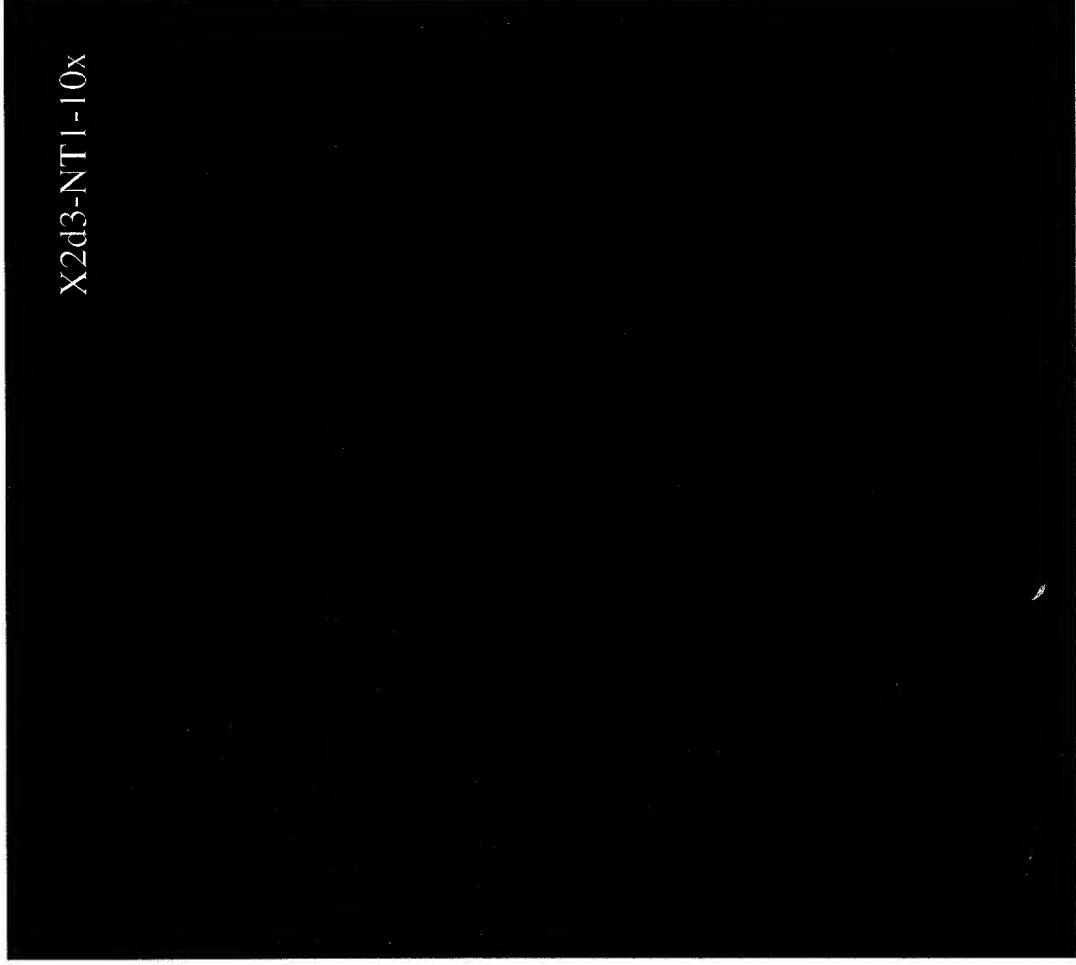
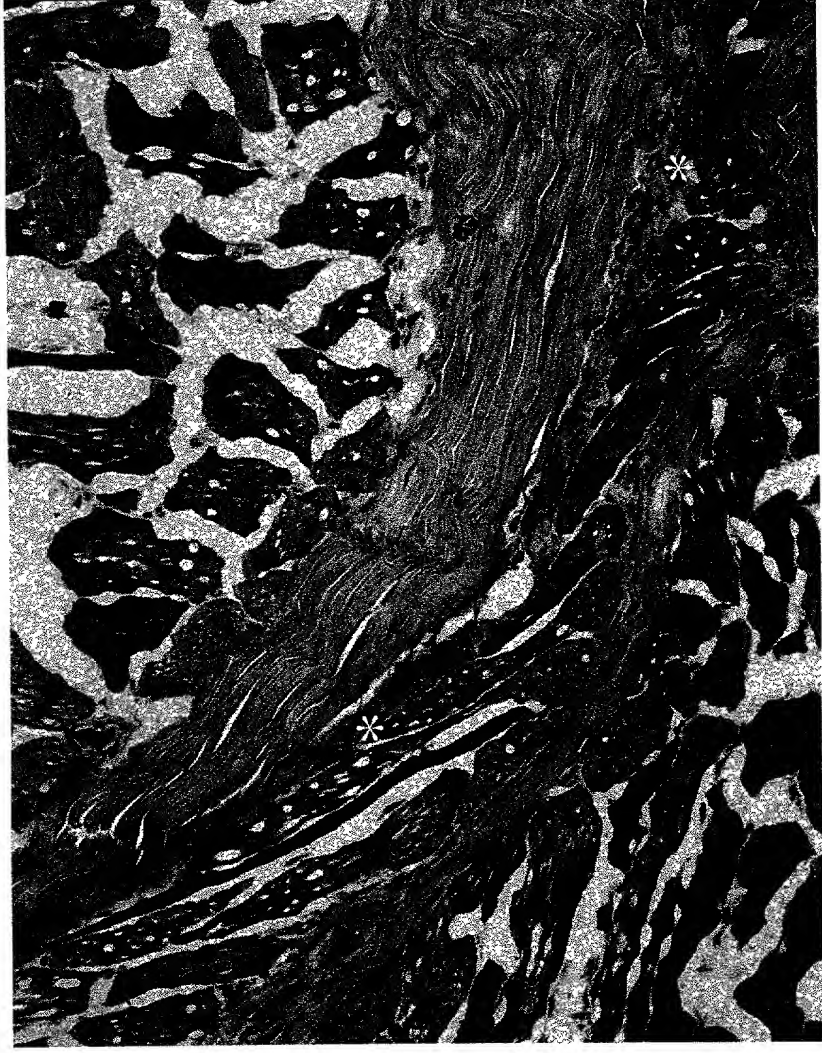


Fig.1b

Human MSCs in Mouse Muscle: Day-7

(H&E)



Dil labelled cells +
Sybr-green staining
of all nuclei

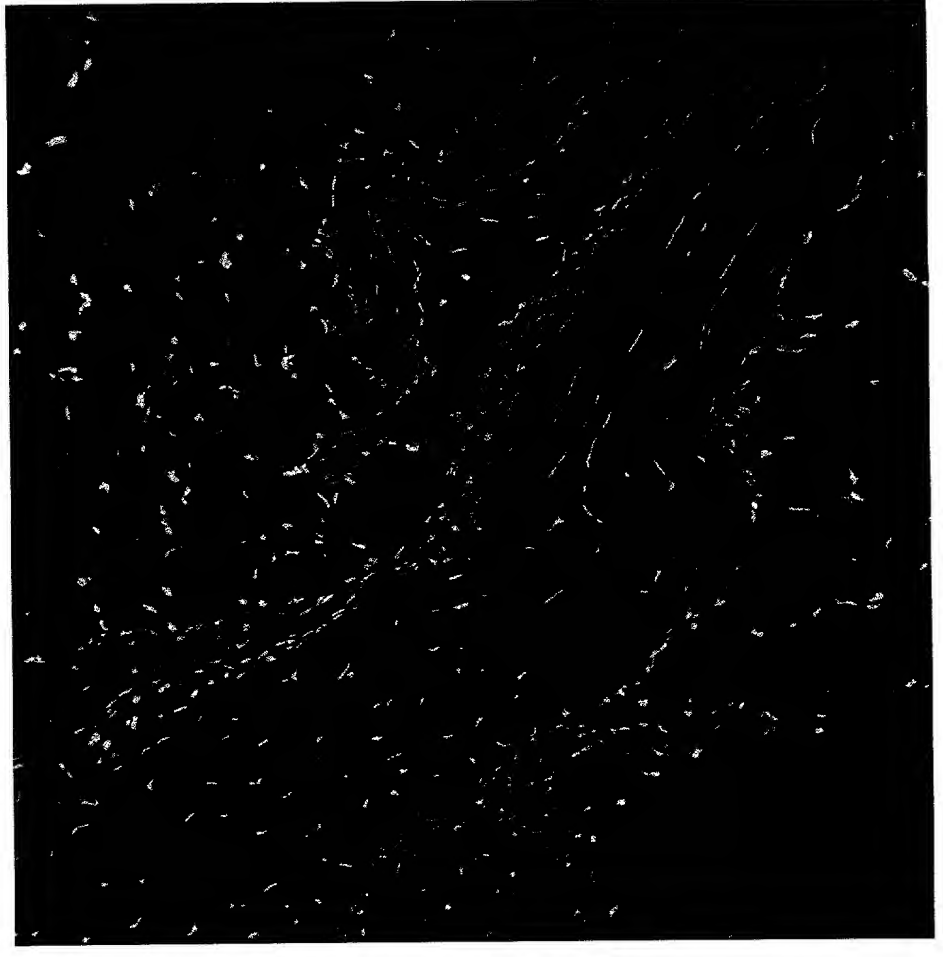


Fig.1c Human MSCs in Mouse Muscle: Day-7

(H&E, 20X)

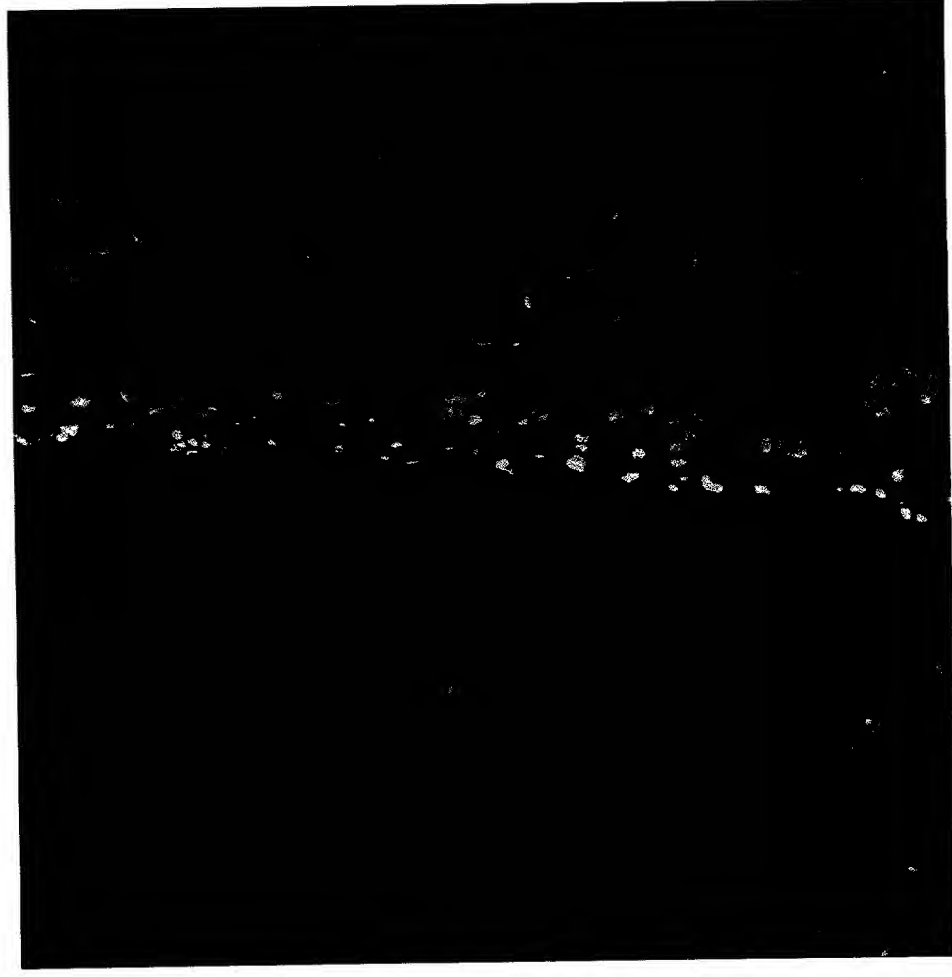
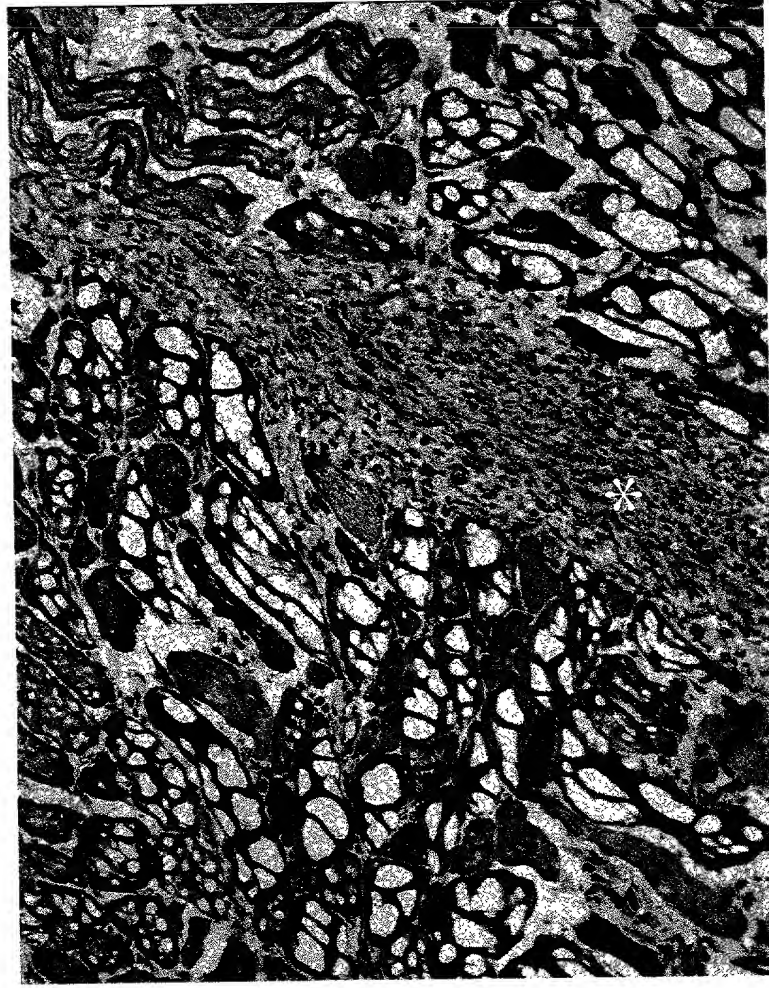
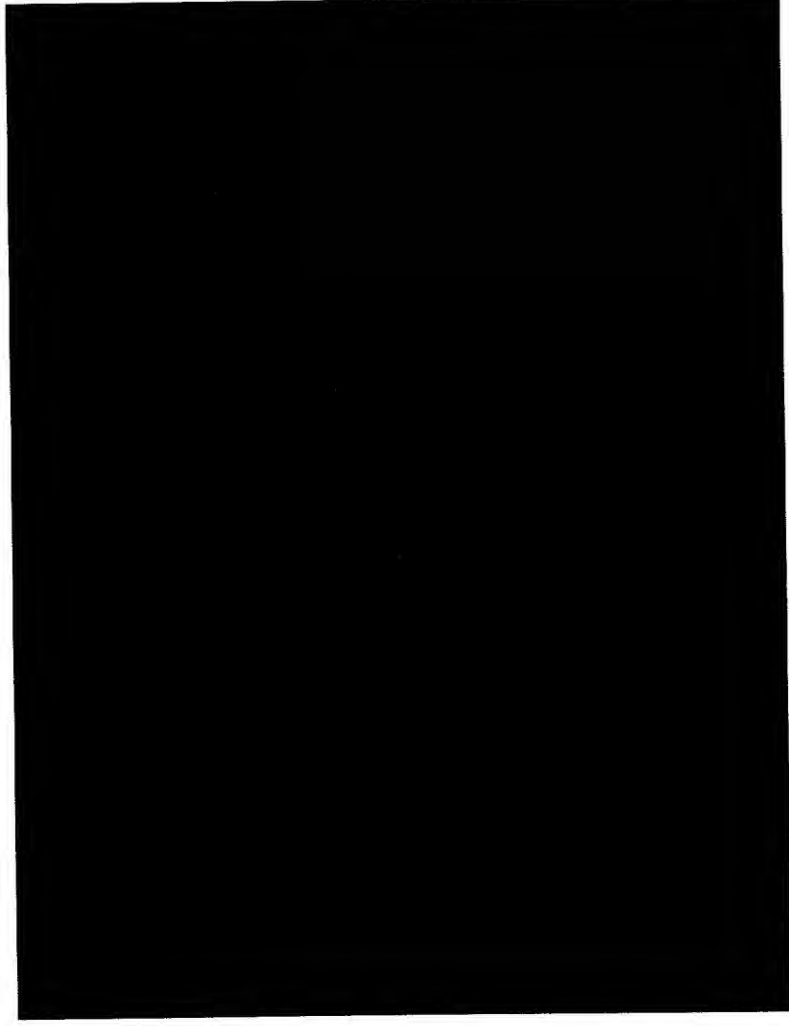
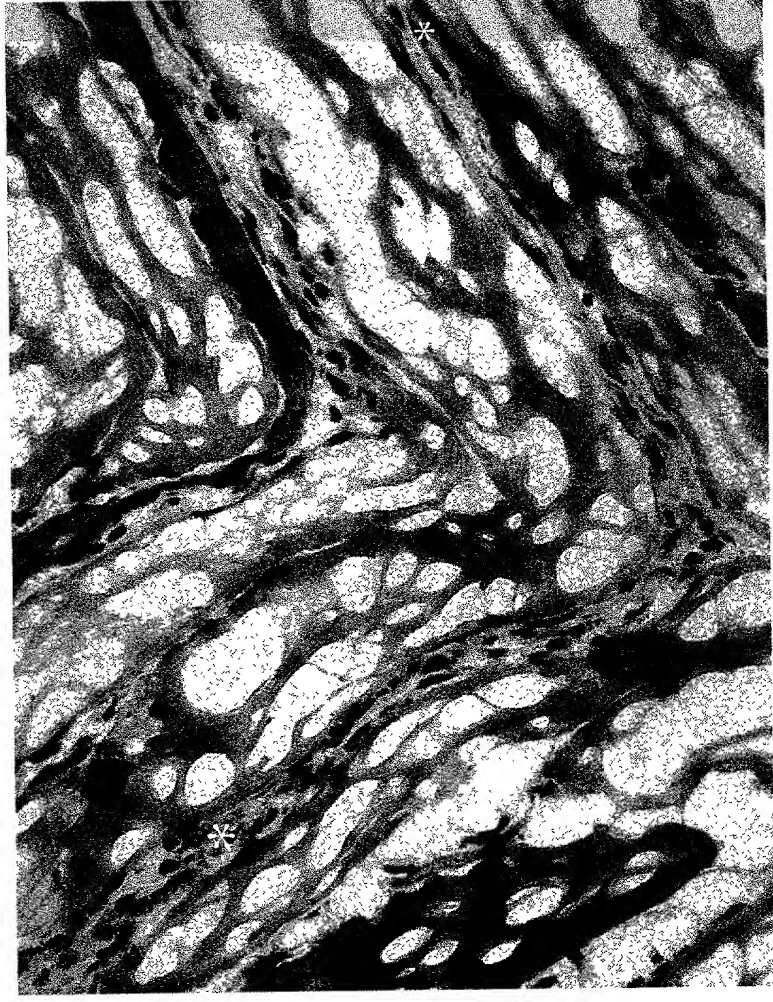


Fig.1d Human MSCs in Mouse Muscle: Day 14

Fluorescent Field, 10X

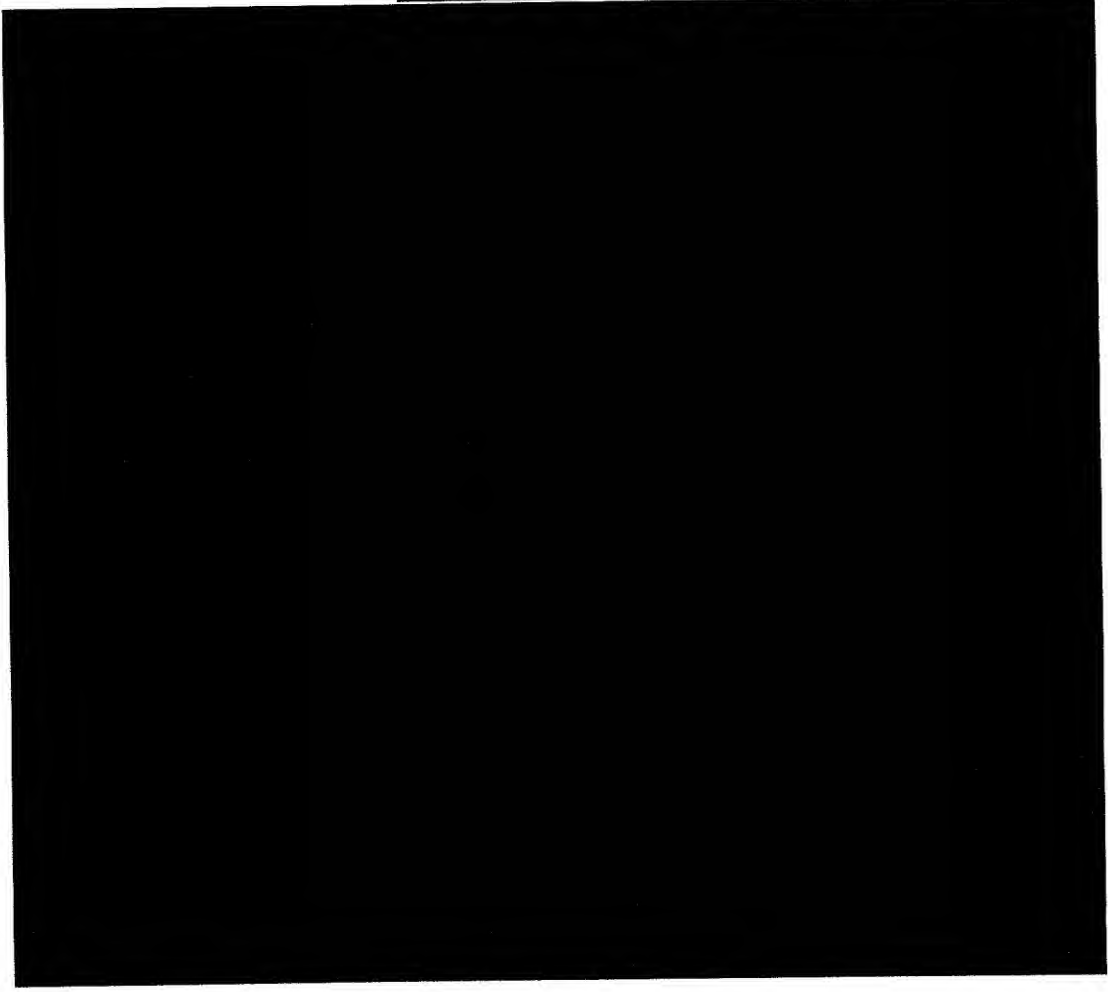


H&E 40X



**Fig. 1e Dil -Positive α -Gala Transduced hMSCs in
Immuno-competent mouse muscle**

d29



d43

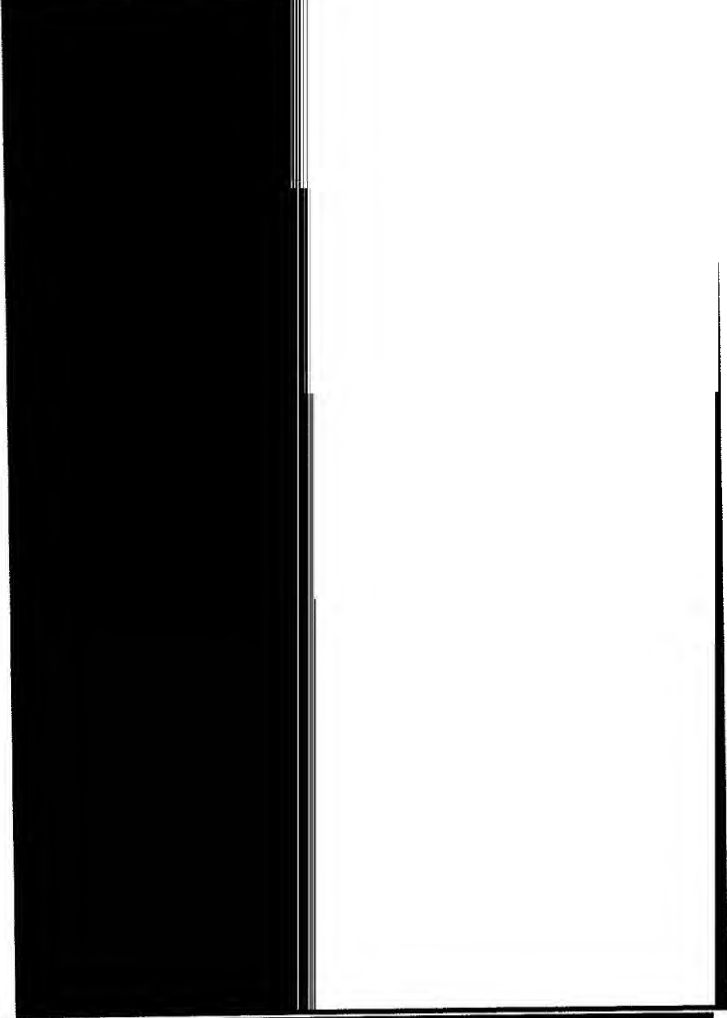


Fig.1f

Human α GalA-Transduced MSCs in Mouse Muscle: Day-57

10X

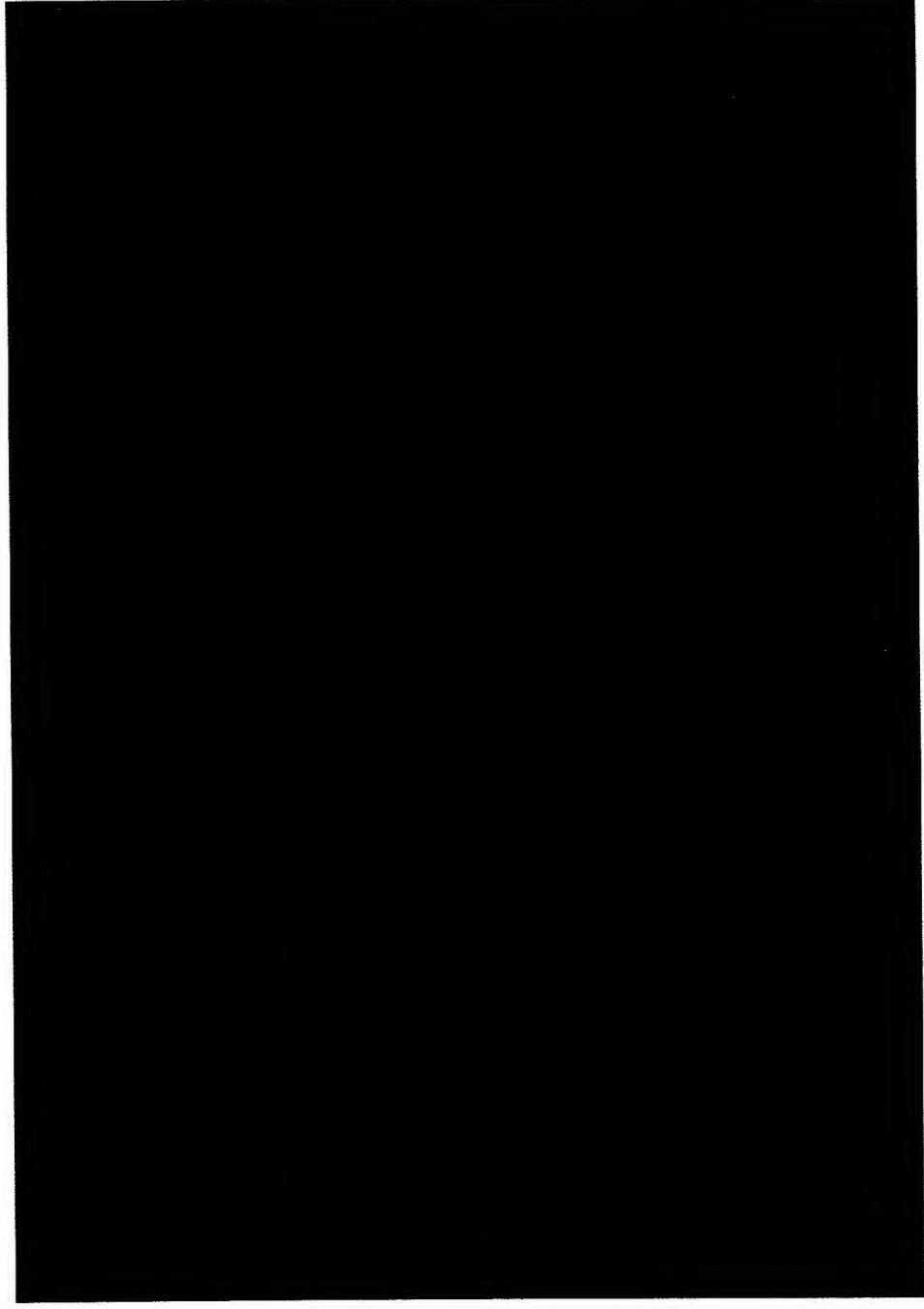


Figure-2

D3: IM injection of Dil-labeled PBMCs

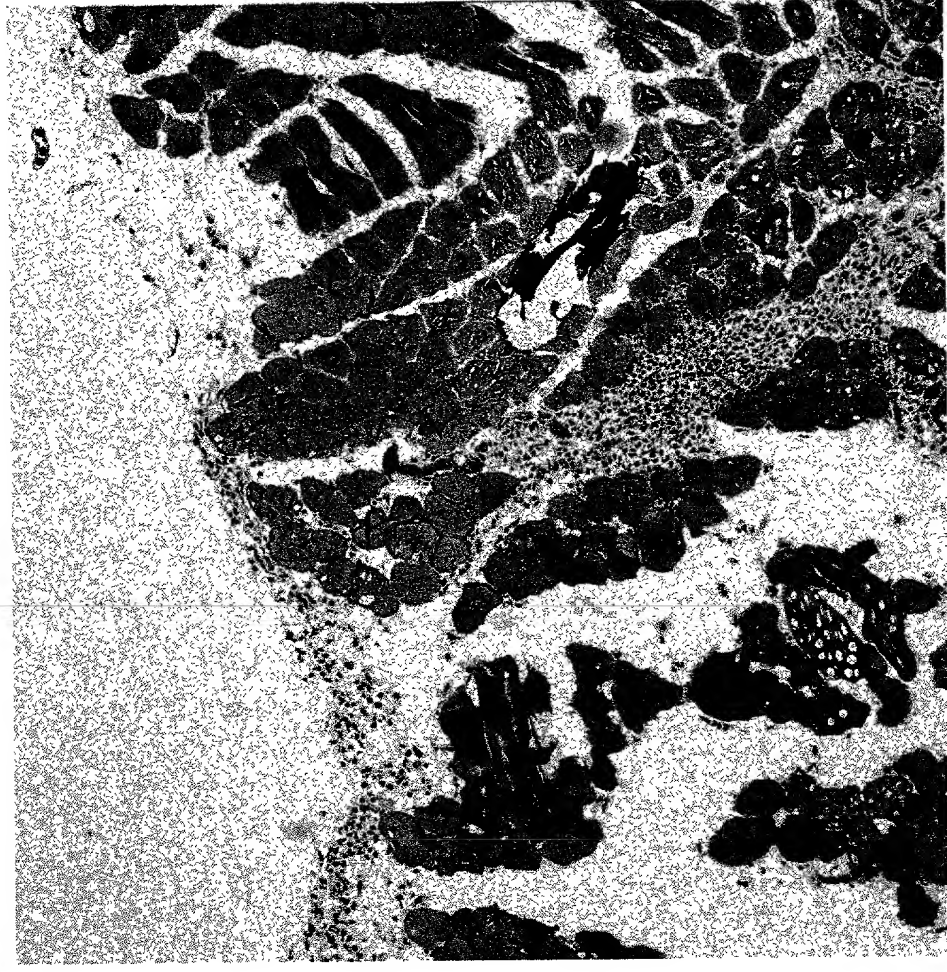
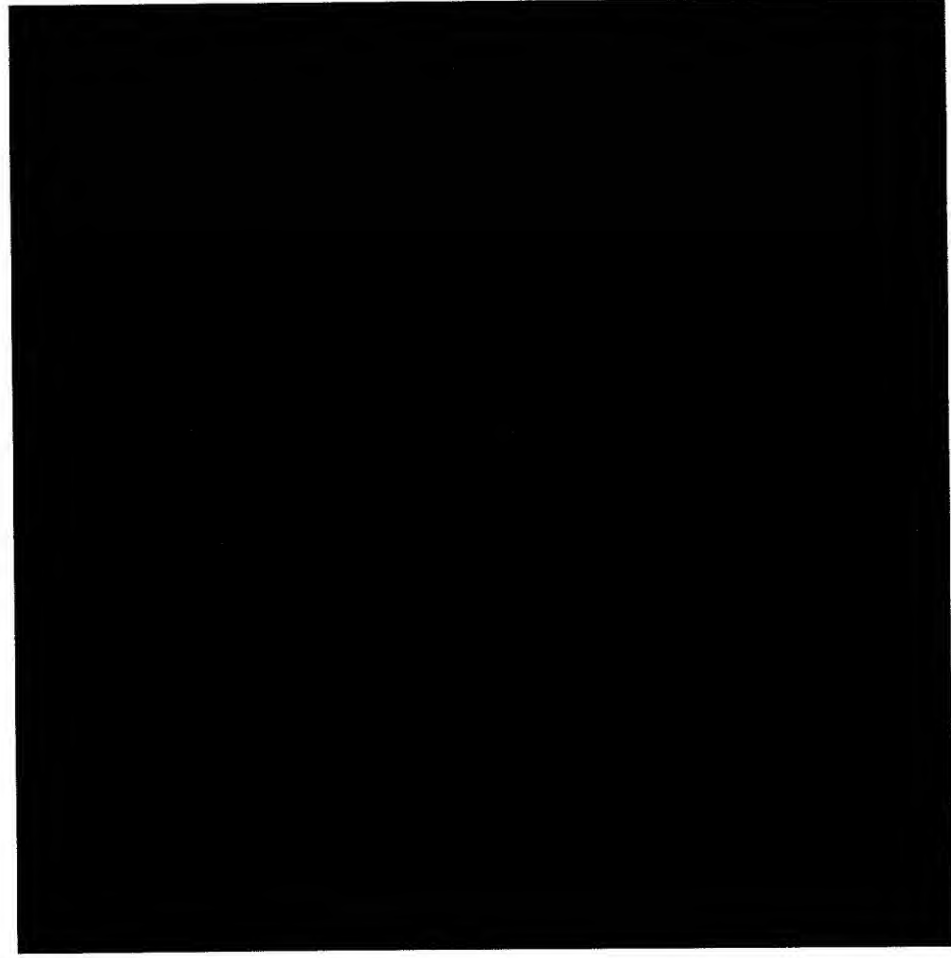
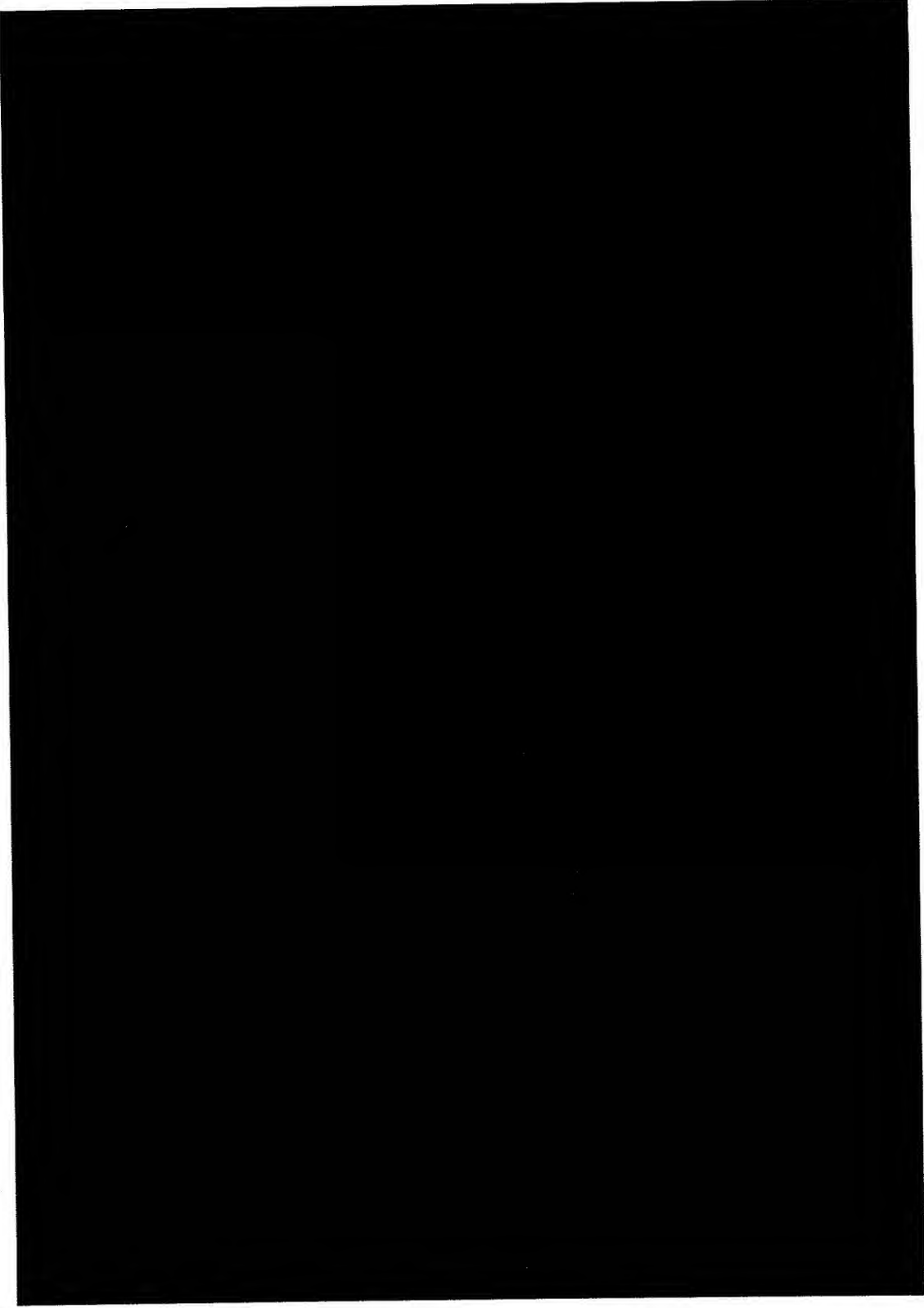


Figure-3

D3 IM injection of saline



Fig. 4 6-week DAPI labeled hMSCs in Mouse Muscle



Red and Blue Florescence Channels merged

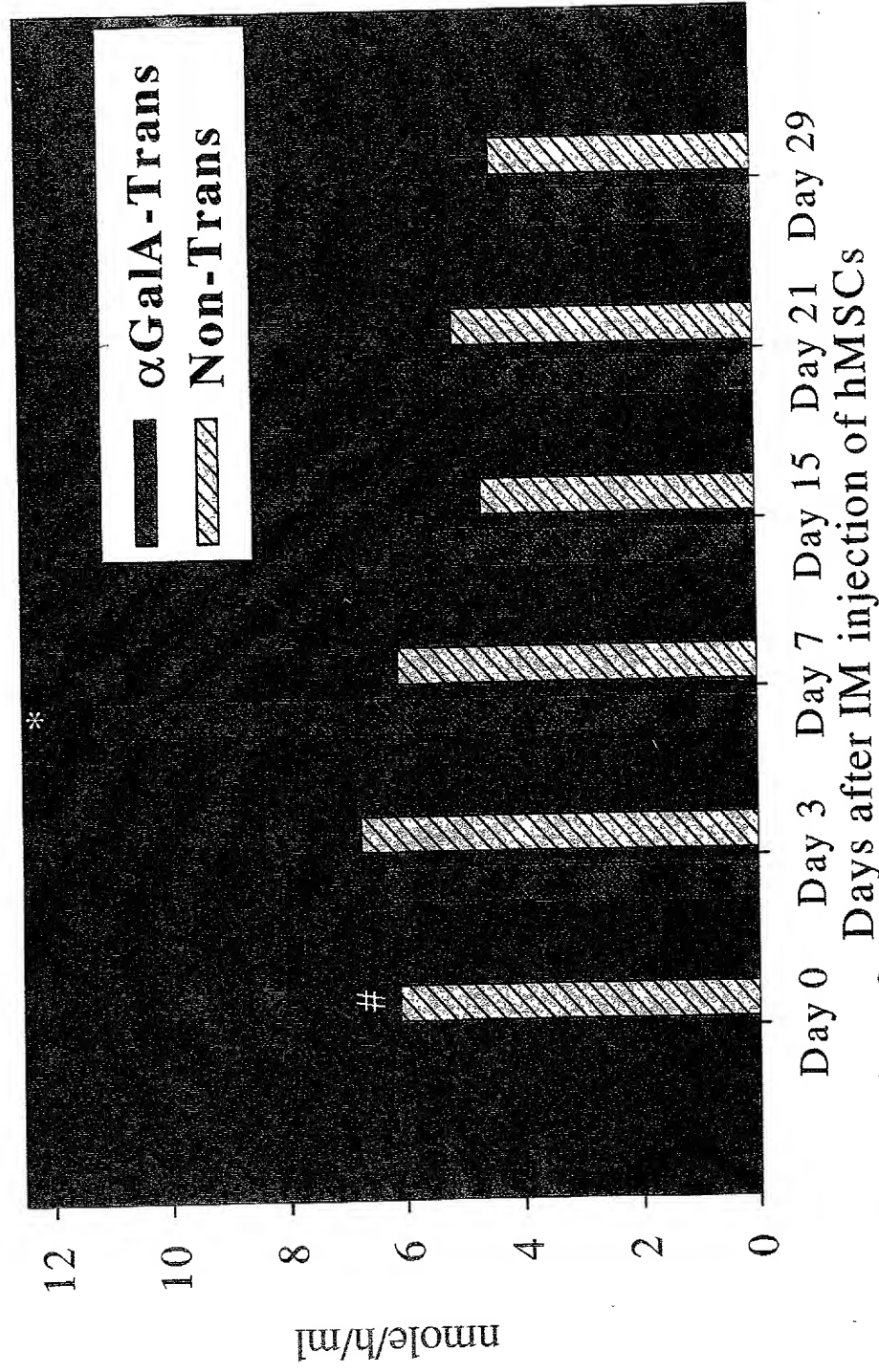
Fig.5

α GalA Immunohostain of Muscle from Mouse
injected IM with α GalA-hMSCs: 14 days



Figure 6

α -GAL activity in Plasma of Mice IM-injected with α -GAL-Transduced hMSCs



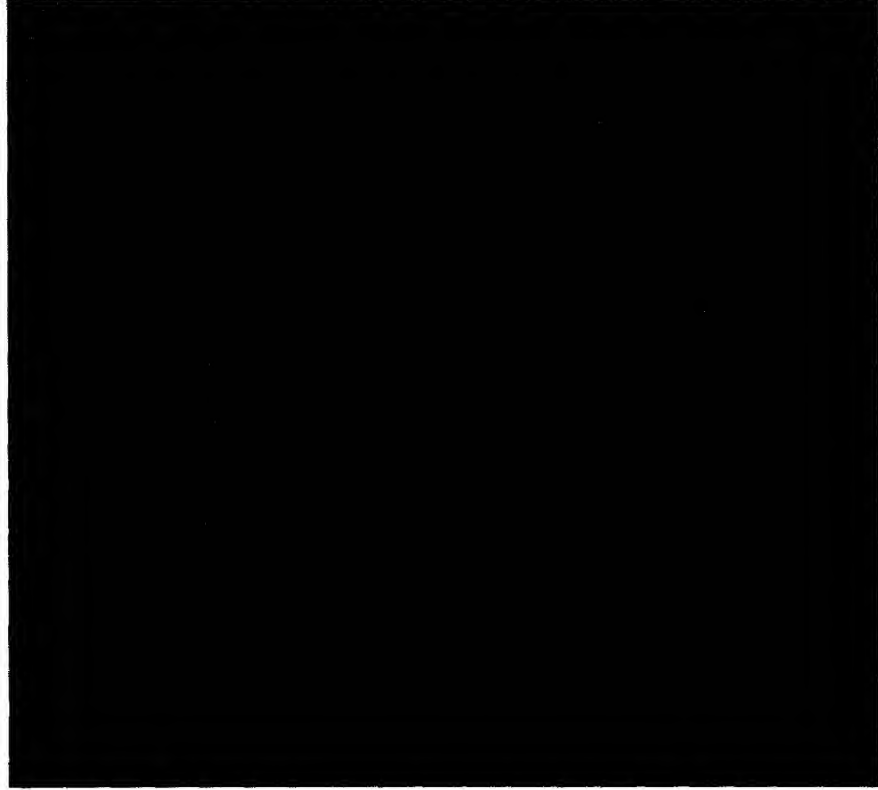
#: No MSCs, * n=2,

Fabry Mice plasma: 0.5 nmole/h/ml, PNAS 97:7515

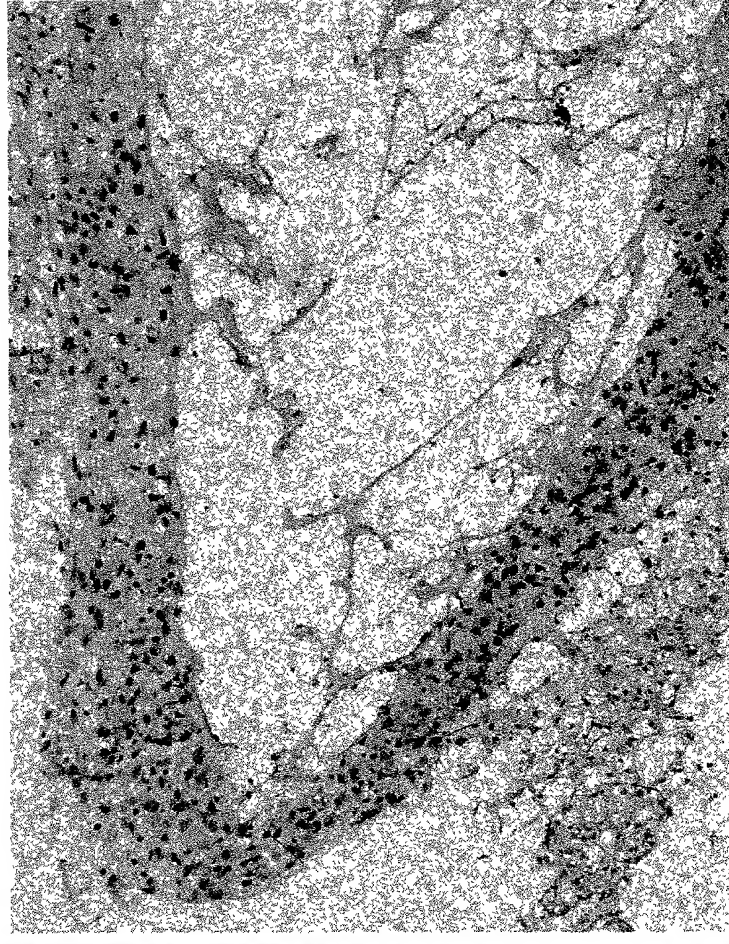
Figure 7a

Human-specific Alu sequence staining of hMSCs injected into Immuno-competent mouse muscle

Dil-positive cells



Alu-sequence positive cells

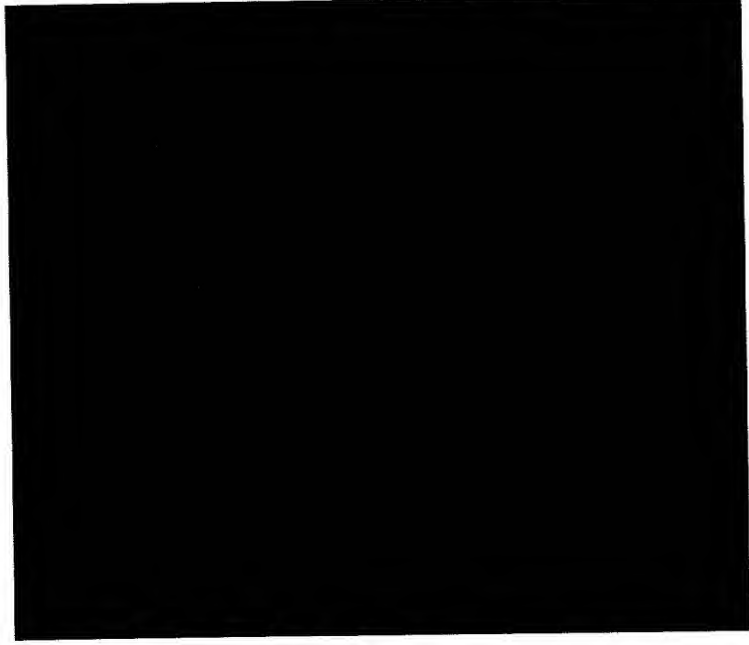


Human MSCs in Mouse Muscle: 43 days

H & E



Dil Positive Cells



Alu-Positive cells

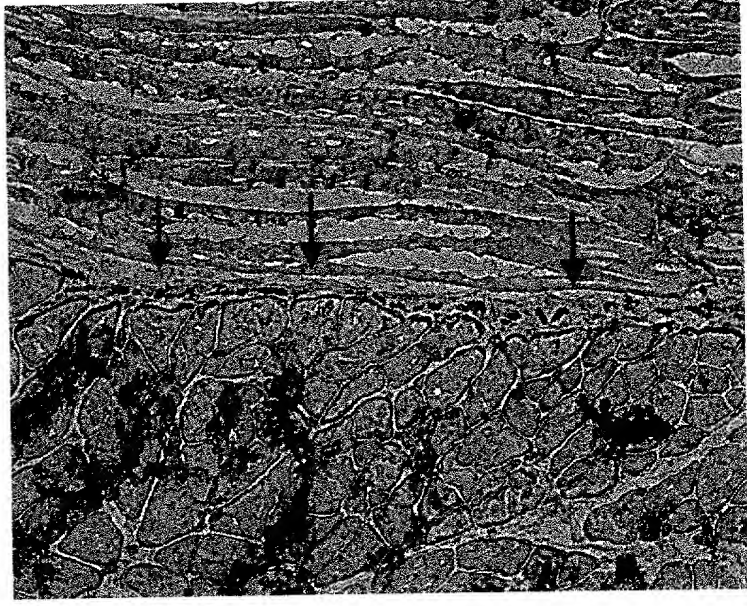


Figure 7c

Day 57 Dil/Alu

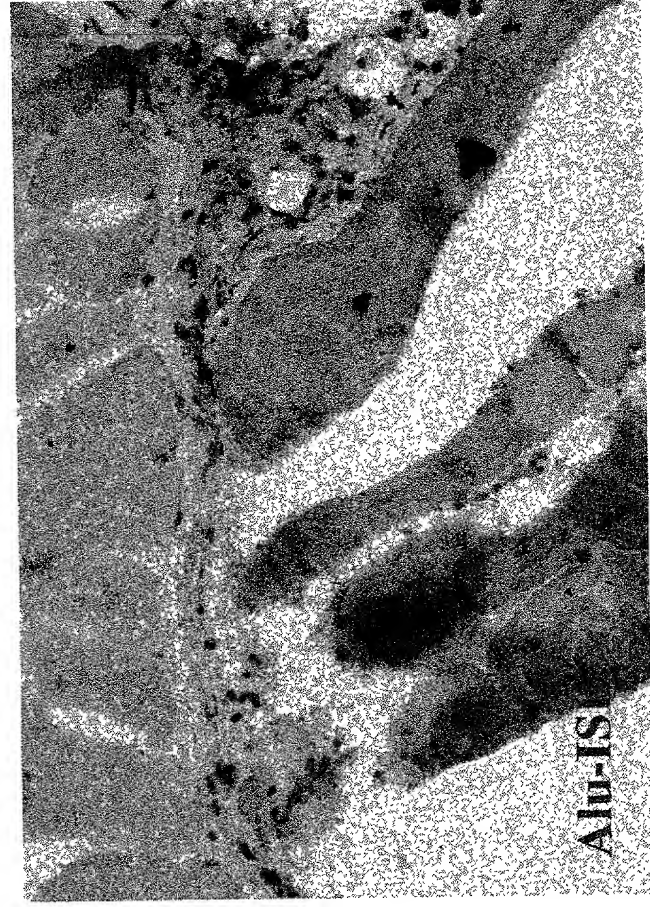
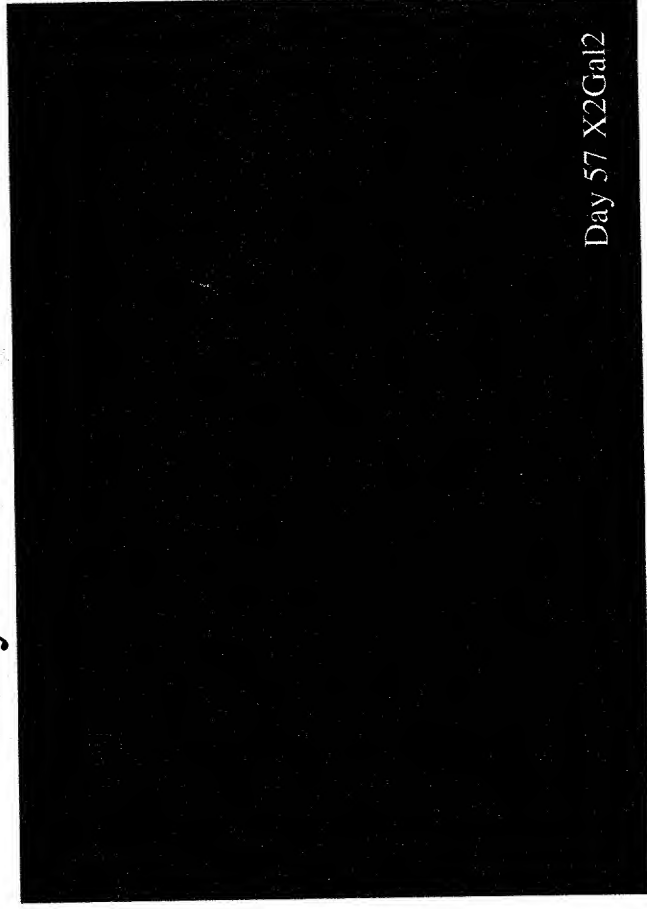
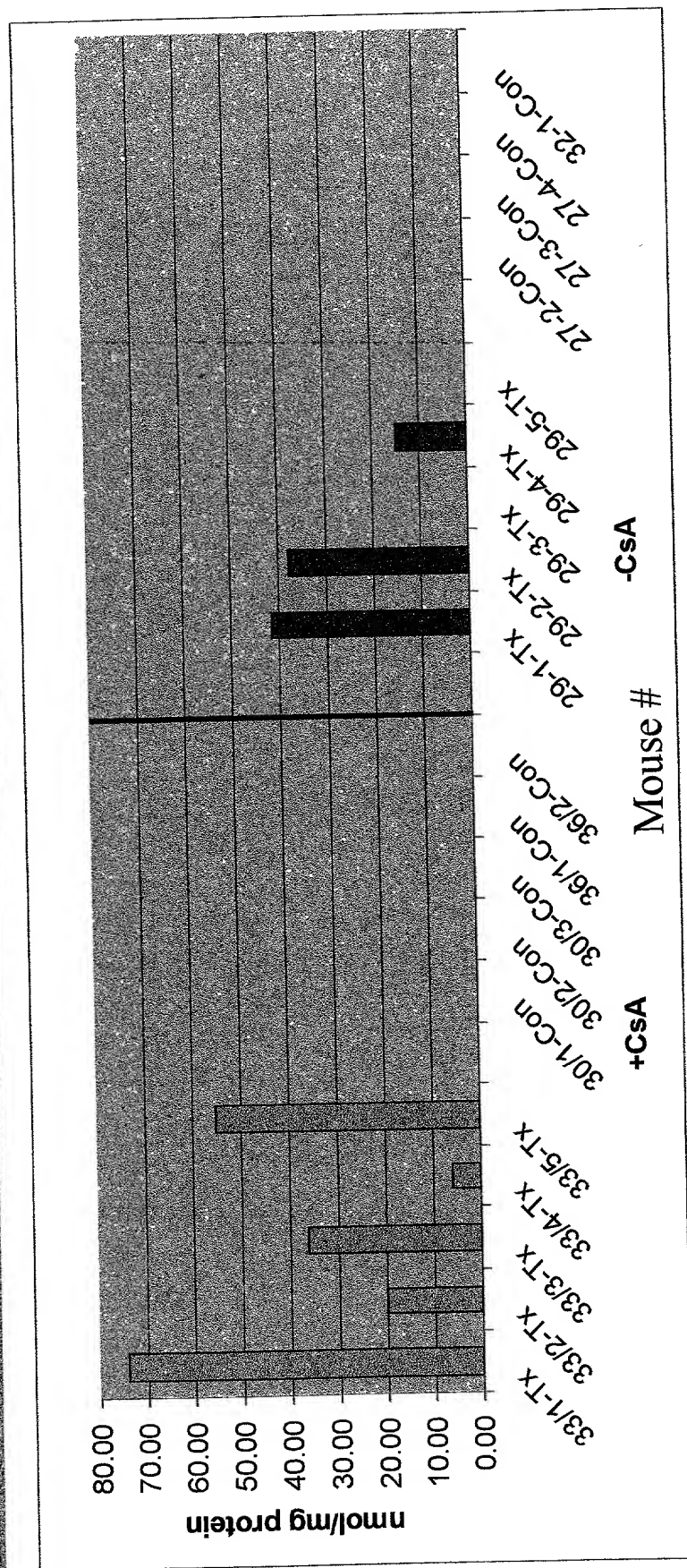


Figure-8a

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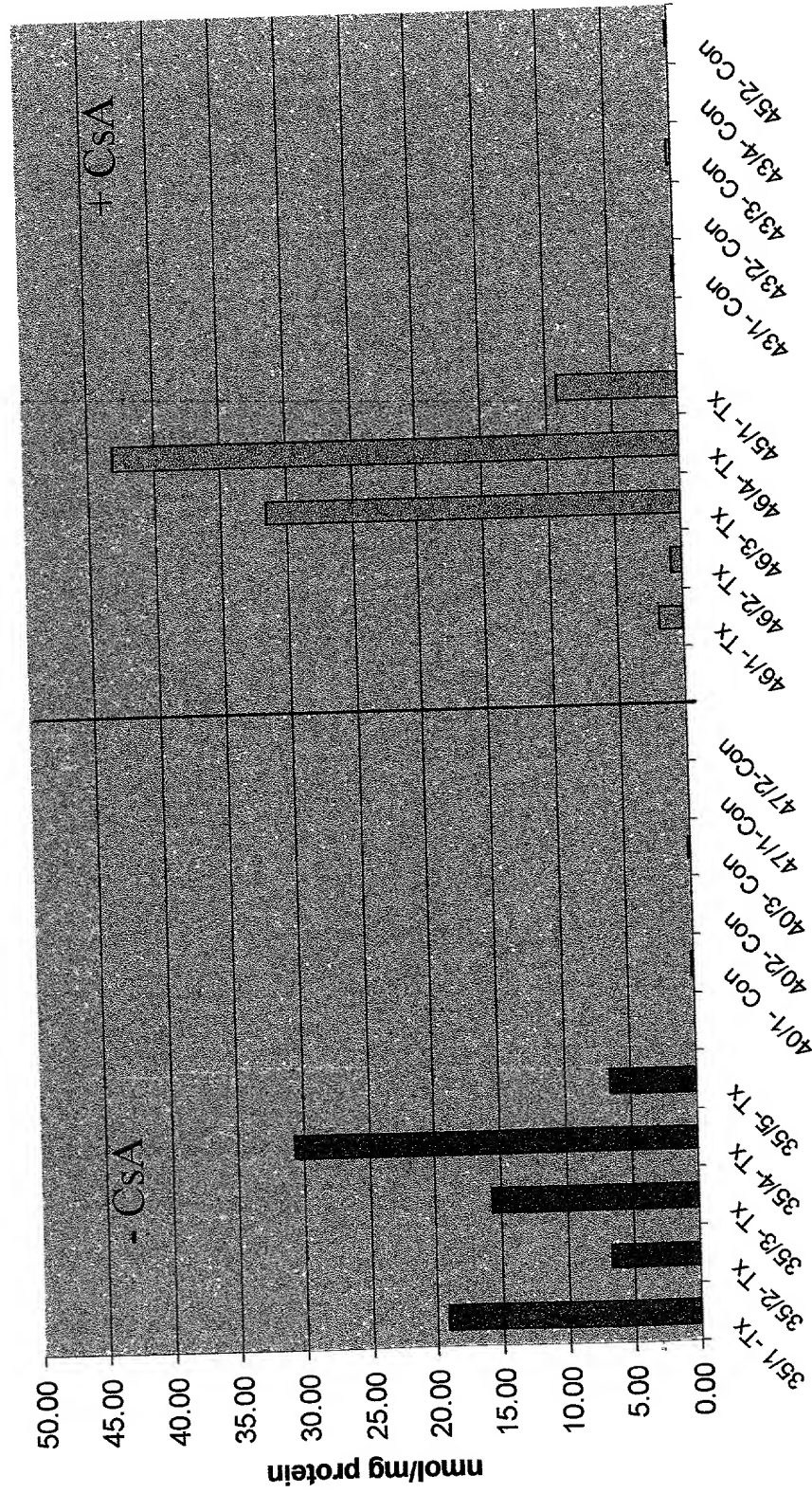
α GalA Activity in Muscles of Fabry-KO mice: 14 days after IM injection of α GalA-Transduced hMSCs



Tx = α GalA Transduced cells injected into muscle

Con = Vehicle alone injected into muscle

Figure-8b **αGAL Activity in Muscles of Fabry-KO mice:**
28 days after IM injection of αGalA-Transduced hMSCs



Tx = αGalA Transduced cells injected into muscle

Con = Vehicle alone injected into muscle

Figure-9a

α GalA Immunostain of Liver from KO Mouse
injected IP with α GalA-hMSCs on Cultisphers :

14 days

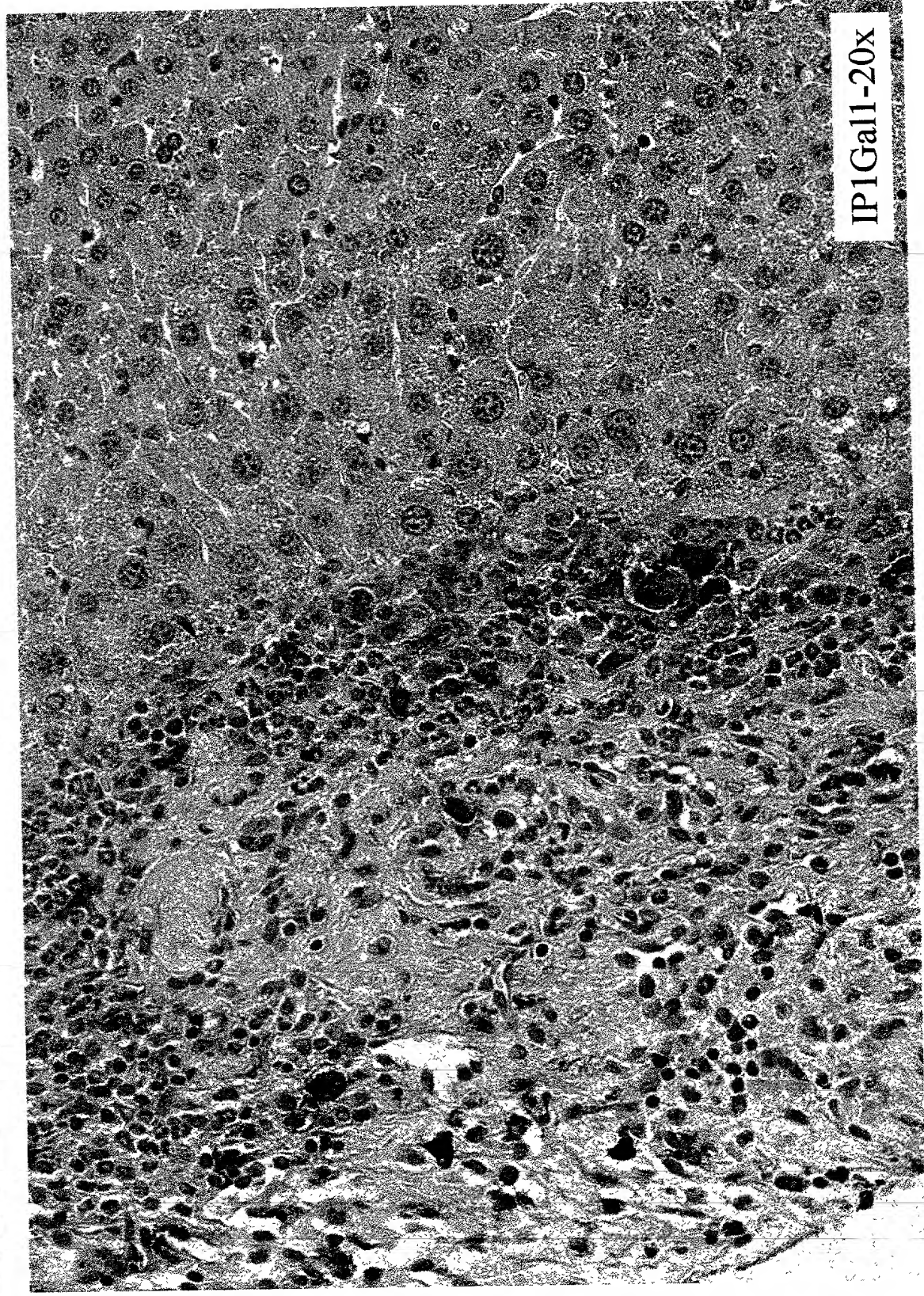


Figure-4b

FOF1^{+/+} Liver from KO Mouse
 α GalA Immunostain of Liver from KO Mouse
injected IP with α GalA-hMSCs on Cultispheres :

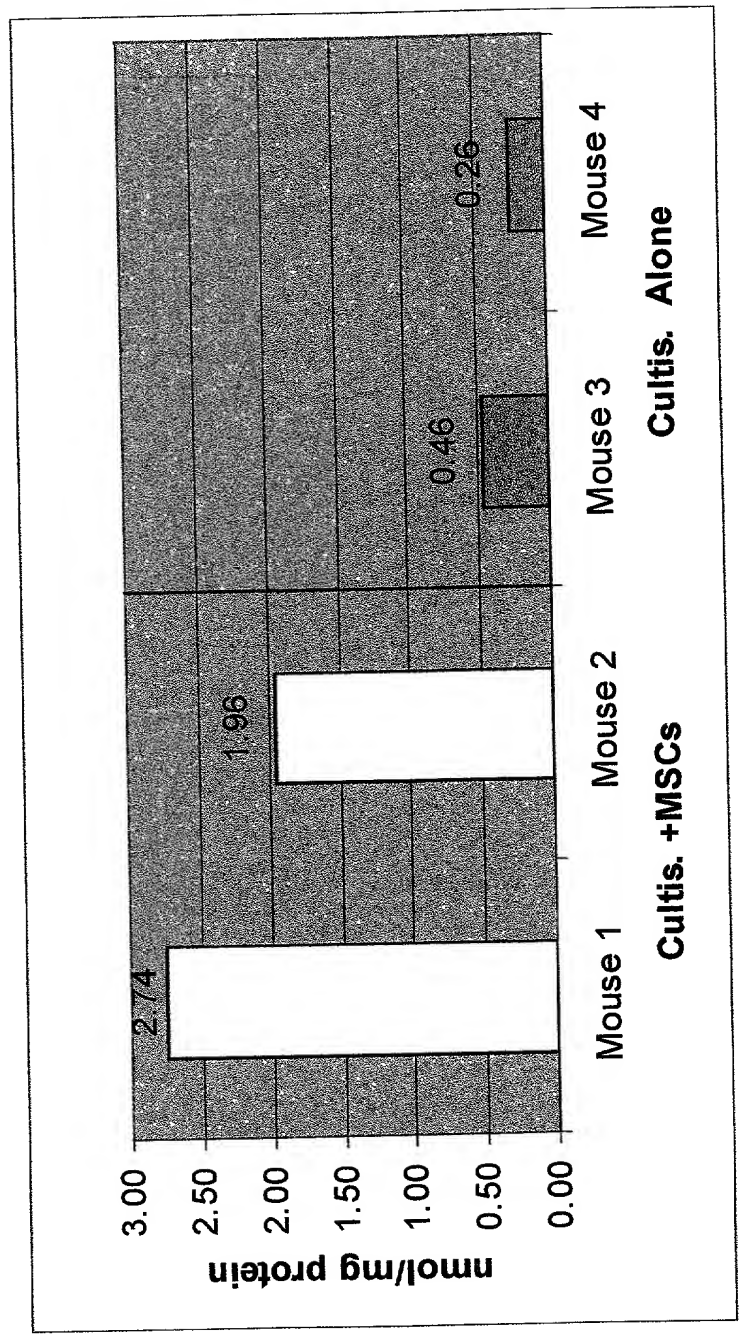
14 days



IP2Gal1a-20x

Intra-Peritoneal Delivery of α GalA-Transduced hMSCs to Fabry Knock-Out Mice

α GalA Enzyme



Increase in α GalA is 5% of normal mouse liver activity (41 nmole/mg)